

# DRAMATIC SUBPERIOSTEAL BONE FORMATION FOLLOWING PHYSEAL INJURY IN PATIENTS WITH MYELOMENINGOCELE

Joseph G. Khoury M.D.; Jose A. Morcuende M.D., Ph.D.

## ABSTRACT

**We present five cases of dramatic subperiosteal bone formation following fracture of the distal femoral physis in children with myelomeningocele. These fractures often go unrecognized due to lack of displacement and/or pain. Children present with a swollen, warm extremity and often systemic signs of inflammation, but rarely any pain or history of trauma. The dramatic radiographic appearance can cause a diagnostic dilemma. We present these cases in order to familiarize the clinician with the oftentimes dramatic appearance of this injury.**

## INTRODUCTION

Lower extremity fractures in children with myelomeningocele are common. They occur in 20% of patients and usually before the age of 9 years.<sup>8</sup> The presentation is often elusive with swelling and warmth of the lower extremity and frequently with a temperature, elevated leukocyte count and sedimentation rate.<sup>4,12,13</sup> There is rarely any associated pain or history of trauma.<sup>3,7,8,9,11</sup> Initially, the work-up may consist of evaluation for other, more common sources of infection. The warm edematous limb frequently becomes suspect for cellulitis and/or osteomyelitis and even sarcoma in light of the other clinical findings. Treatment and further workup will proceed along this course unless the pattern of healing is recognized on the radiograph. Failure to make the diagnosis initially was common in the cases we reviewed.

---

Department of Orthopaedic Surgery  
University of Iowa Health Care  
Iowa City, Iowa, U.S.A.

### Address correspondence and reprint requests to:

Jose Morcuende M.D.  
Department of Orthopaedic Surgery  
University of Iowa Health Care  
200 Hawkins Dr.  
Iowa City, IA. 52242

No financial support of any kind was received in the preparation of this manuscript.

## CASES

### Case 1

This nearly five year old female with a T12 myelomeningocele spent 6 weeks in a spica cast after bilateral anterior hip releases. One week after cast removal she presented with painless left thigh edema. A radiograph revealed a Salter Harris type II fracture of the distal femoral physis (Figure 1A). She was placed in a knee immobilizer. Follow up films three weeks later (Figures 1B-C) show extensive subperiosteal bone formation encompassing nearly the entire femoral shaft.

### Case 2

This 11 year old male with L1 myelomeningocele presents to clinic before his regularly scheduled routine visit because his caretaker noticed right distal thigh swelling, redness, warmth and fevers. He was admitted to the hospital with the presumptive diagnosis of cellulitis and treated with antibiotics. When he failed to respond in a few days, radiographs of the limb were obtained to rule out osteomyelitis (Figures 2A-B). The radiographs revealed a Salter Harris II injury to the distal femoral physis with extensive periosteal elevation along the entire shaft of the femur. Treatment consisted of a brace and activity modification for six weeks.

### Case 3

This two year old male with L1 myelomeningocele spent five weeks in a hip spica cast following bilateral anterior hip releases and bilateral tendoachilles lengthening. A few days after cast removal, he was noted to have a fever at home. He was brought to the pediatrician for a work-up. At that time, he was noted to have edema, warmth and erythema of both distal thighs. Radiographs were obtained (Figures 3A-C). There was no pain or history of trauma.

### Case 4

This two and a half year old female with L4 myelomeningocele spent eight weeks in a hip spica cast after bilateral proximal femoral osteotomies. Three days after cast removal she was brought in by her mother with edema and warmth of the thigh. Infection was initially suspected. Radiographs revealed a distal femoral



**Figure 1A**



**Figure 1B**



**Figure 1C**

**Figure 1.** (A) AP radiograph of the femur immediately after injury. (B, C) AP radiograph of the proximal and distal femur three weeks after injury.



**Figure 2A**



**Figure 2B**

**Figure 2.** (A, B) AP and lateral radiographs of the femur several days after presentation.



**Figure 3A**



**Figure 3B**



**Figure 3C**

**Figure 3.** (A) AP radiograph of both femurs. (B, C) Lateral radiographs of right and left femurs.



**Figure 4A**

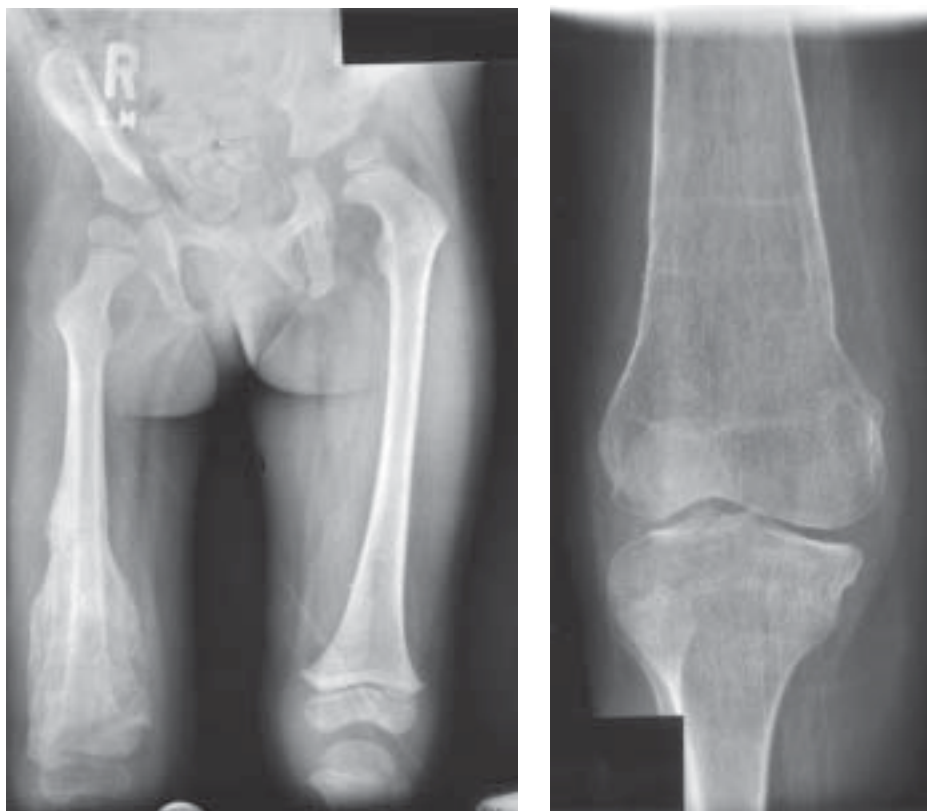


**Figure 4B**



**Figure 4C**

**Figure 4.** (A,B) AP and lateral radiographs of the left femur upon presentation. (C) Lateral radiograph of the left femur 15 months after the injury.



**Figure 5.** (A) AP radiograph of both femurs. (B) AP radiograph of the knee taken 13 years after the injury.

physeal injury (Figures 4A-B). She was immobilized again in plaster for 3 weeks. Radiographs after removal showed exuberant callus formation. Days later she was noted to have a proximal tibial physeal injury as well. Note the Erlenmeyer flask deformity at her final follow up 15 months later (Figure 4C).

### **Case 5**

This three year old female with a high level myelomeningocele suffered a right distal femoral physeal injury three days after coming out of a hip spica cast. Radiographs two months after healing (Figure 5A). Persistent Erlenmeyer flask deformity 13 years later (Figure 5B).

### **DISCUSSION**

Most lower extremity fractures in myelodysplastic children are diaphyseal or metaphyseal and although there is rarely much displacement, are relatively easy to recognize on plain radiograph. Nine percent of fractures are through the physis and are more likely due to repetitive stresses in children with some mobility potential.<sup>7</sup> Younger patients with myelomeningocele undergo more frequent surgical procedures and subse-

quent post-operative immobilization. This adds to their underlying osteopenia and susceptibility to fracture when the immobilization is discontinued.<sup>1,4,8</sup>

Physeal fractures are more difficult to identify on plain radiograph. They are usually minimally displaced due to the thick periosteal sleeve. Frequently all that is seen acutely is widening of the physis. However, most physeal injuries are discovered in the subacute period when callus has begun to form. Callus is frequently abundant and may be due to delayed presentation and lack of immobilization.<sup>11</sup> While diaphyseal and metaphyseal fractures heal readily with minimal treatment,<sup>4,7,8</sup> physeal injuries require more aggressive, prolonged treatment in order to avoid growth arrest and deformity.<sup>7</sup> However, if they are detected early, predictable and reliable healing can be achieved with simple avoidance of physical therapy and strenuous activity while using their usual

braces for a period of one month.<sup>9</sup>

The radiographic appearance of a subacute fracture can range from simple abundance of local callus to exuberant, subperiosteal bone formation along the entire length of the affected bone. Misdiagnosis is frequent at this stage. Metaphyseal flaring can simulate numerous disorders including fibrous dysplasia, Gaucher's disease, heavy metal poisoning and various metaphyseal dysplasias.<sup>5</sup> The exuberant and disorganized callus formation can simulate rickets, scurvy or syphilis.<sup>4</sup> Also, these patients frequently present with local and systemic signs of inflammation which, when combined with the radiographic appearance often leads to the diagnosis of osteomyelitis or osteosarcoma.<sup>4,8,12,13</sup> While callus can often be extensive in sensate children as well, it remodels as the bone begins to experience the normal stresses of weightbearing after healing. The changes in myelomeningocele can persist for many years.<sup>5</sup> We presented two cases with sufficient follow-up to demonstrate the Erlenmeyer flask appearance of the metaphysis years after the injury.

The exact etiology of this phenomenon is strictly speculative. The degree of periosteal stripping and subsequent bleeding into the potential space can give the

radiographic appearance of pseudotumor of hemophilia.<sup>2</sup> The flaccid muscle tissue may allow more periosteal stripping and a larger hematoma to form before tamponade occurs.<sup>5</sup>

The dramatic radiographic appearance that often follows these simple physeal injuries can pose a diagnostic challenge to the untrained observer. The amount of bone formation far from the injury is impressive. Careful inspection of the physis and correlation with the clinical presentation can aid in the diagnosis of this interesting condition.

### CONCLUSION

Children with myelodysplasia are at high risk for lower extremity fractures due to osteopenia and lack of sensation. Younger patients are at especially high risk after a period of immobilization following surgery. These injuries can often go unrecognized for a period of time. Patients frequently present with local and occasionally systemic signs suggestive of infection. It is important to recognize the clinical and radiographic features of this entity in order to avoid unnecessary work-up and provide appropriate treatment.

### REFERENCES

1. **Anschuetz RH, Freehafer AA, Shaffer JW, Dixon, Jr MS.** Severe Fracture Complications in Myelodysplasia. *J Pediatr Orthop.* Vol. 4, No. 1, 1984.
2. **Dirschl DR, Greene WB.** Pseudotumor of the Distal Part of the Femur in a Patient Who Had Myelomeningocele. *J Bone Joint Surg Am* 1992;74:935-8.
3. **Golding, C.** Museum Pages III. Spina Bifida and Epiphyseal Displacement. *J Bone Joint Surg Br* 1960;42-B:387-9.
4. **Handelsman JE.** Proceedings and Reports of Councils and Associations. *J Bone Joint Surg Br* 1972;54:381.
5. **Hyre HM, Stelling CB.** Radiographic Appearance of Healed Extremity Fractures in Children with Spinal Cord Lesions. *Skeletal Radiol* (1989) 18:189-192.
6. **Katz JF.** Spontaneous Fractures in Paraplegic Children. *J Bone Joint Surg Am* 1953;35:220-6.
7. **Kumar SJ, Cowell HR, Townsend P.** Physeal, Metaphyseal, and Diaphyseal Injuries of the Lower Extremities in Children with Myelomeningocele. *J Pediatr Orthop.* Vol. 4, No. 1, 1984.
8. **Lock TR, Aronson DD.** Fractures in Patients Who Have Myelomeningocele. *J Bone Joint Surg Am* 1989;71:1153-7.
9. **Roberts JA, Bennet GC, MacKenzie JR.** Physeal Widening in Children with Myelomeningocele. *J Bone Joint Surg Br* 1989;71-B:30-32.
10. **Rodgers WB, Schwend RM, Jaramillo D, Kasser JR, Emans JB.** Chronic Physeal Fractures in Myelodysplasia: Magnetic Resonance Analysis, Histologic Description, Treatment, and Outcome. *J Pediatr Orthop.* Vol. 17, No. 5, 1997.
11. **Schneider R, Goldman AB, Bohne WHO.** Neuropathic Injuries to the Lower Extremities in Children. *Radiology* 128:713-718, September 1978.
12. **Souter FE.** Spina Bifida and Epiphyseal Displacement. *J Bone Joint Surg Br* 1962;44-B:106-9.
13. **Townsend PF, Cowell HR, Steg NL.** Lower Extremity Fractures Simulating Infection in Myelomeningocele. *CORR* Number 144, October 1979.